

# Radiological findings and the clinical importance of megacalycosis

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**Objective:** To describe the radiological findings and the clinical importance of megacalycosis.

**Materials and methods:** On the basis of a case report and literature review, diagnostic criteria and clinical significance of megacalycosis are presented.

**Result:** Megacalycosis is mostly asymptomatic and is usually discovered either accidentally or as a result of its complications, such as stone formation, flank pain, hematuria, infection, and fever. The renal pelvis, infundibulum, and ureter are not dilated. Calyces have a semilunar configuration rather than the conventional triangular or conical form. The tip of each pyramid is flat, and the calyces possess neither fornix nor papillae impressions. The number of calyces is increased compared to the healthy condition, typically from 20–25. The renal parenchyma has a normal width but with a slight narrowing of the renal medulla. The kidney exhibits normal function, in particular with respect to its ability to concentrate the urine.

**Conclusion:** Megacalycosis is a rare, usually unilateral dilatation of the kidney calyces in the presence of a normal, undilated renal pelvis and ureter. Its pathological significance lies in the occurrence of complications.

**Keywords:** megacalycosis, megacalycose, calyx dilatation, stone formation

## Introduction

Puigvert was the first to describe the status of a nonobstructive dilatation of the calyces in the absence of a dilated renal pelvis and ureter as megacalycosis.<sup>1</sup> This condition usually concerns only one kidney and should be distinguished from hydronephrosis due to the lack of renal pelvis and ureter dilatation and the presence of a normal kidney parenchyma.<sup>2,3</sup>

Although megacalycosis is not hereditary, it has been associated with the Schinzel–Giedion syndrome, a rare autosomal recessive disorder that is characterized by coarse facial features, midface retraction, hypertrichosis, multiple skeletal anomalies, and bilateral megacalyces.<sup>4,5</sup> In these cases, megacalycosis was associated with proteinuria and uremia. The condition is also thought to have an autosomal recessive mode of inheritance, as evidenced by the existence of three children from a marriage between relatives (cousins) who exhibited bilateral megacalyces.<sup>6</sup>

Although the exact pathogenesis of the megacalycosis remains unclear, there is no evidence of a causal relationship of this condition with vesicoureteral reflux, papillary necrosis, or chronic infection. A possible congenital underdevelopment of the renal pyramids is believed to be responsible for the dilatation of the calyces.<sup>3</sup>

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Johnston suggested a temporary fetal and spontaneously regressed pyeloureteral obstruction in a 9-year-old girl as the cause of her unilateral megacalycosis and spoke of a burnt-out obstruction. However, in contrast to a true megacalycosis, the kidney of this patient exhibited a slight renal pelvis dilatation.<sup>7</sup>

Kimche and Lask considered that dilatation of the calyces could be attributed to the absence of peristalsis of the calyces, similar to achalasia.<sup>3</sup> This theory was supported by the observations of megacalyces in patients with congenital Hirschsprung's disease.<sup>8</sup>

Yet another hypothesis postulates a tissue weakness in the renal calyces, which occurs secondary to megacalyces through high physiological pressure in the renal pelvis.<sup>9</sup>

Megacalycosis is mostly asymptomatic and is usually discovered accidentally or by associated complications, such as stone formation, flank pain, hematuria, infection, and fever.

## Case presentation

The protocol for the project was approved by the Ethics Committee of the Democritus University of Thrace, and the patient consented to the publication of the data. A 45-year-old woman was referred by her gynecologist to clear up a "hydronephrosis" of the left kidney. The supposed hydronephrosis was accidentally discovered by an ultrasound examination because of uncharacteristic right-sided abdominal pain probably as a result of adnexitis. Urine examination was completely normal, and the performed intravenous pyelography showed the characteristic signs of a megacalycosis of the left kidney. A possible obstruction of the left kidney could be excluded.

## Discussion

Megacalycosis is a rare, usually unilateral dilatation of the kidney calyces in the presence of a normal, undilated renal pelvis and ureter.

In a narrower sense, megacalycosis is not a disease but rather a condition, and as such the term "megacalycoses" is recommended instead megacalycosis.

Approximately 100 cases with unilateral or bilateral megacalyces are described in the literature.<sup>1,3,6,10-12</sup> Part of them was presented with urinary tract infection or kidney colic. For most cases, however, especially in children, congenital hydronephrosis was falsely supposed. Megacalycosis coexisted with a primary megaureter in five of these cases.<sup>10</sup>

## Imaging

Imaging studies such as intravenous pyelogram and computed tomography urography have revealed that renal calyces have



**Figure 1** Intravenous urography of kidney with megacalycosis.

a semilunar configuration rather than the classic triangular or conical form (Figure 1).

The tip of each pyramid is flat, and the calyces possess neither fornix nor papillae impressions. Furthermore, the number of calyces is increased compared to the normal condition, from typically<sup>7-9</sup> to 20–25.

The renal pelvis, infundibulum, and ureter are not dilated in megacalycosis, thus differentiating it from kidney congestion. In addition, the renal parenchyma shows a normal width but with a slight narrowing of the renal medulla. Angiographic studies have revealed normal interlobular blood vessels that curve slightly around the dilated calyces.<sup>11</sup>

The kidney function is normal in megacalycosis, in particular with respect to its ability to concentrate the urine, unless it is damaged by the frequent occurrence of complications. Disturbance of the urine outflow as a result of the megacalyces can produce a tendency toward stone formation in patients.

## Clinical observations

Megacalycosis is generally discovered either accidentally or as a result of associated complications, including stone

formation, hematuria, infection, or flank pain. Therapeutic management of the condition is limited to the treatment of its complications; operative management, such as pyeloplasty, is obsolete.

## Diagnostic procedure

Intravenous urography in combination with renal ultrasound, computed tomography, or magnet resonance urography is recommended for the diagnosis. In children, voiding cystourethrography should be performed to rule out vesicorenal reflux. Endoscopic measures, such as retrograde ureterography and ureterorenoscopy, are not required for diagnosis and can lead to unnecessary infections; therefore, this technique should be avoided.

## Conclusion

Megacalycosis should not be confused with hydronephrosis, since this would lead to unnecessary operative measures. Renal function is not impaired in megacalycosis, but due to the complication of urine stasis in the megacalyces, there exists a tendency toward stone formation and infection in patients. The therapeutic management of this condition involves the treatment of complications that have already been presented and the prevention of lithiasis and infection by ensuring an adequate intake of fluids.

## Disclosure

The authors report no conflicts of interest in this work.

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